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The Fontan Procedure for Pulmonary Atresia With Intact Ventricular Septum: Operative and Late Results

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Objectives. **The goals of the study were to evaluate the operative and late mortality associated with the Fontan procedure in patients with pulmonary atresia and an intact ventricular septum and to obtain follow-up information on the current clinical status of surviving patients.**

Background. **Between 1979 and October 1, 1995, 40 patients with the anomaly had a nonfenestrated Fontan procedure performed at the Mayo Clinic. Because there are no previously published reports involving a series of this size in which the Fontan approach was used for this condition, a review of patient outcomes was thought to be of value.**

Methods. **The medical records of the 40 patients were reviewed retrospectively, and 34 were determined to be alive. The status of the survivors as of late 1995 was then ascertained by direct examination, questionnaire or telephone follow-up.**

Results. **There were three operative deaths and three late deaths. The current ages of the 34 survivors ranged from 4 to 30 years (median 13). Thirty-three of the 34 survivors were thought to be in New York Heart Association functional class I or II, and all but three of these patients, of school age or older, were either full-time students or working full time. The three adults who were not employed thought they were capable of working but were not doing so because of socioeconomic reasons. More than half of the patients were not receiving cardiovascular medications.**

Conclusions. **These overall gratifying early and late results encourage continued application of this operation for appropriately selected patients with this complex congenital cardiovascular anomaly.**

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Pulmonary atresia with an intact ventricular septum is an uncommon but not rare congenital heart malformation. The size and morphology of the tricuspid valve and right ventricle in this lesion can vary widely, and fistulous connections between the right ventricle and coronary arterial system are often found, particularly in patients with a very hypoplastic right ventricle (1–3). Ultimately, the size of the tricuspid valve and right ventricle, and occasionally the presence of coronary artery obstructive lesions in association with right ventricle to coronary artery fistulas, will dictate whether patients will be candidates for a two-ventricle repair or whether a less corrective procedure such as the Fontan operation will be the most definitive surgical option (4).

Several recent publications (2–5) have discussed the optimal neonatal surgical management of patients with pulmonary atresia and an intact ventricular septum—the goal being to eventually achieve a two-ventricle repair. However, a significant proportion of these patients—those with markedly hypoplastic tricuspid valves and right ventricles, and occasionally obstructive coronary artery lesions—will not become candidates for a two-ventricle repair, regardless of what procedure is chosen for their neonatal surgical management. For this group, the Fontan procedure, which does achieve the goal of separating the systemic and pulmonary circulations and thereby eliminating arterial hypoxemia and ventricular volume overloading, becomes the most definitive surgical procedure available.

Methods

Patients. Between 1979 and October 1, 1995, 40 patients with pulmonary atresia and an intact ventricular septum had a nonfenestrated Fontan procedure performed at the Mayo Clinic. These patients represent ~4.5% of the total of 890 patients at our institution who had nonfenestrated Fontan procedures performed from its inception in 1973 through October 1, 1995.

Age at the time of the Fontan procedure ranged from 1 year, 10 months to 21 years (median 6 years) (Table 1). The 40 patients had undergone a total of 74 previous palliative cardiovascular surgical procedures (mostly at other institutions) before their Fontan procedure (Table 2). Only one patient had not undergone previous surgical palliation; 16 had undergone one procedure; 12 patients, two procedures; 10

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Table 1. Age at Operation in 40 Patients Who Had a Fontan Operation for Pulmonary Atresia With Intact Ventricular Septum, 1979 to October 1, 1995

Age at Operation (yr)*	No. of Patients
0–2	3
3–5	16
6–10	13
11–15	5
>15	3

*Range 1 year, 10 months to 21 years (median 6 years).

patients, three procedures; and one patient, four previous cardiovascular operations.

Tricuspid valve diameter, as determined echocardiographically or angiocardiographically, ranged from 11% to 63% of that expected for a patient's body surface area, excluding one patient with a normal-size tricuspid valve and right ventricle who had multiple right ventricle to coronary artery fistulas and coronary artery obstructive lesions ("right ventricular-dependent coronary circulation") (1,3). Right ventricular volume, determined echocardiographically or angiocardiographically, varied between 8% and 55% of that expected, excluding the patient with a normal-size tricuspid valve and right ventricle.

Five patients had angiocardiographically demonstrated right ventricle to coronary artery fistulas, but only one (the child with the normal-size tricuspid valve and right ventricle) had obstructive coronary artery lesions producing right ventricular dependence for a portion of the coronary circulation.

Surgical management. Surgical management of the tricuspid valve at the time of the Fontan procedure for these 40 patients is shown in Table 3. In 17 patients the valve was left open to receive systemic venous blood. In 10 more recent cases in which the lateral tunnel technique (6) was used to route systemic venous blood to the pulmonary arteries, the tricuspid valve was left patent to receive pulmonary venous blood. Seven patients, who all had the operation between 1979 and 1984, had patch or suture closure of the valve. In six patients with previous right ventricular outflow tract reconstructions to establish right ventricle to pulmonary artery continuity, the hypoplastic tricuspid valve and right ventricle were left in the circulation between the right atrial appendage and the main pulmonary artery.

Table 2. Previous Procedures in 40 Patients Who Had a Fontan Operation for Pulmonary Atresia With Intact Ventricular Septum

Previous Palliative Procedure	No. of Patients
Blalock-Taussig shunt (classic or modified)	33
Waterston shunt	11
Central Gore-Tex shunt	10
Pulmonary valvotomy or RVOT reconstruction	7
Classic right Glenn anastomosis	5
Potts shunt	2
Bidirectional cavopulmonary anastomosis	1
Other	5
Total	74

RVOT = right ventricular outflow tract.

Table 3. Management of Tricuspid Valve in 40 Patients Who Had a Fontan Operation for Pulmonary Atresia With Intact Ventricular Septum

Tricuspid Valve Management	No. of Patients
Left patent to receive systemic venous blood	17
Left patent to receive pulmonary venous blood (lateral tunnel technique)	10
Patch or suture closure*	7
Hypoplastic tricuspid valve and right ventricle left in the right atrial appendage to pulmonary artery circulation	6
Total	40

*All between 1979 and 1984.

The types of connections used to direct systemic venous blood to the pulmonary arteries are shown in Table 4. Twenty-eight patients had anastomosis of the right atrium or right atrial appendage directly to the main or right pulmonary artery, and in four of these patients a previous classic right Glenn anastomosis was left intact. Six patients had a direct right atrium to main pulmonary artery anastomosis combined with an end to end classic right Glenn anastomosis performed at the time of their Fontan procedure to direct superior vena caval blood to the right pulmonary artery. This was necessitated by right pulmonary artery distortion and hypoplasia caused by a previous Waterston anastomosis in four patients and by a previous right Blalock-Taussig anastomosis in two others. This was thought to be the best way to manage this problem, rather than by attempting an extensive angioplasty of the hypoplastic proximal right pulmonary artery. As mentioned previously, six patients with previous right ventricular outflow tract reconstruction had a right atrium to pulmonary artery

Table 4. Connections Used to Direct Systemic Venous Blood in 40 Patients Who Had a Fontan Operation for Pulmonary Atresia With Intact Ventricular Septum

Type of Connection	No. of Patients
Right atrium to main or right pulmonary artery*	28
Right atrium to main and left pulmonary artery with superior vena cava to right pulmonary artery†	6
Right atrium to main pulmonary artery anastomosis leaving the hypoplastic tricuspid valve and right ventricle in the circulation‡	4
Right atrium to main and left pulmonary artery anastomosis, leaving the hypoplastic tricuspid valve and right ventricle in the circulation, with superior vena cava to right pulmonary artery§	2

*Four previous right Glenn anastomoses were left intact. †Necessitated by right pulmonary artery distortion due to previous Waterston anastomosis (n = 4) and right Blalock-Taussig anastomosis (n = 2). ‡One previous right Glenn anastomosis left intact. §Necessitated by right pulmonary artery distortion due to previous Waterston anastomosis (n = 2).

Table 5. Surgical Results in 40 Patients Who Had a Fontan Operation for Pulmonary Atresia With Intact Ventricular Septum

Result	No. of Patients (%)
Operative death	3* (8%)
Late death	3† (8%)
Total	6

*One each in 1979, 1984 and 1989. †Two sudden and unexpected deaths at 2½ and 8 years after the Fontan operation. One death due to chronic protein-losing enteropathy and terminal sepsis 8 years after the Fontan operation.

anastomosis, which left the hypoplastic right ventricle and tricuspid valve in the circulation. In one of these patients, a previous classic right Glenn anastomosis was left intact, and in two of these patients an end to end classic Glenn anastomosis was performed, again to manage the problem of severe proximal right pulmonary artery distortion and hypoplasia caused by a previous Waterston shunt.

Results

There were three operative deaths (Table 5), one each in 1979, 1984 and 1989; the operative mortality rate was 8%. Three late deaths occurred (a rate of 8%) at 2½ years, 8 years and 8 years postoperatively. The first two late deaths were sudden and unexpected in patients doing clinically well and with unremarkable results of postmortem examinations; these were presumably due to a dysrhythmia. The other patient died 8 years after the Fontan procedure from complications of chronic protein-losing enteropathy (7); sepsis was the terminal event.

At the time of this report, the ages of the 34 surviving patients ranged from 4 to 30 years (median 13); five patients were young adults between 18 and 25 years old, and five patients were older than 25 years.

The current functional status of all 34 surviving patients was ascertained by examination, questionnaire or telephone follow-up. The duration of postoperative follow-up ranged from 6 months to 13 years (median 6 years). Thirty-three patients were in good or excellent condition and in New York Heart Association functional class I or class II, and all but three, 6 years or older, either were working full time or were full-time students. The three adult patients who were not working full time felt capable of doing so; their unemployment was due to socioeconomic factors. One patient had had a heart transplantation performed at another institution 7 years after the Fontan procedure because of gradual myocardial deterioration but was doing well 3 years after the transplantation. One patient was in fair condition (functional class III) and had diminished ventricular function and persistent atrial dysrhythmias.

Nineteen of the survivors were not receiving cardiovascular medications. Four were receiving digoxin (Lanoxin) only. Two were receiving digoxin and an intermittent diuretic, and two were receiving digoxin and an afterload reducer. The patient who had the most recent operation, who was followed up 6 months after the Fontan operation, was taking digoxin, a diuretic, and an afterload reducer. The patient in fair condition was receiving digoxin, diuretics, an afterload reducer and antidysrhythmic medications. Three patients were receiving digoxin and an antidysrhythmic agent because of intermittent atrial dysrhythmias, but they stated that their rhythm problems were well controlled by their medical regimen. One patient was receiving warfarin anticoagulation because of a mild cerebrovascular accident that occurred ~1 year after the Fontan procedure, but this patient was doing well, without neurologic sequelae, 18 months after the cerebrovascular accident. The patient who received the heart transplant was on a standard antirejection protocol.

Discussion

In a recent publication, Bull et al. (3) stated, "In the 1990s pulmonary atresia with intact ventricular septum remains a dismal lesion." They then conceded, however, that the advent in recent years of early neonatal echocardiographic diagnosis and immediate treatment with prostaglandin had improved the survival of patients with this ductus-dependent lesion and that neonatal strategies must be planned with a view toward later, possibly more definitive, surgical treatment.

Neonatal management. Immediate neonatal surgical management involves establishment of a reliable and adequate source of pulmonary blood flow. In patients with only mild to moderate hypoplasia of the tricuspid valve and right ventricle, this is accomplished by right ventricular outflow tract reconstruction to establish right ventricle to pulmonary artery continuity, with or without an accompanying systemic to pulmonary artery shunt. This approach in these patients accomplishes the dual goal of providing adequate pulmonary blood flow while also stimulating additional tricuspid valve and right ventricular growth to maximize chances of a later two-ventricle repair (4).

In patients with severe tricuspid valve hypoplasia and a diminutive right ventricle, however, it has become clear that there is little potential for significant tricuspid valve enlargement and right ventricular growth, even if right ventricle to pulmonary artery continuity is established early (2,4), and so these patients are treated with a neonatal systemic to pulmonary artery shunt only with an eye toward later conversion to the Fontan circulation (8,9). It is this group of patients with severe right heart hypoplasia who are particularly prone to the presence of right ventricle to coronary artery fistulas (4,10), and the "dismal" outlook for these patients described by Bull et al. (3) relates in part to a significant incidence of sudden, presumably dysrhythmic, deaths, possibly due to coronary insufficiency caused by a "steal" phenomenon (11). Obliteration of the hypoplastic ventricular cavity surgically or with catheterization embolization techniques in patients with clear-cut ventricle to coronary artery fistula, but without distal obstructive lesions in the coronary tree, is advocated by some

investigators (12) as a means to eliminate "steal" from the coronary circulation with its secondary potential for sudden death.

Fontan procedure. The unfavorable natural history of congenital heart lesions characterized by a functional single ventricle led to the search for an innovative surgical way to separate the systemic and pulmonary circulations in these malformations. More than 25 years have passed since the initial Fontan operation was performed to achieve this purpose (13), and during this time the procedure has undergone many technical modifications (14) and more definitive criteria for proper selection of patients have evolved (14,15). Nearly 900 nonfenestrated Fontan procedures have been performed at our institution for various forms of the single ventricle since the initial procedure was done here in 1973. Operative mortality in recent years has declined to <5% for the lesions of tricuspid atresia and double-inlet left ventricle and to <10% in the more complex forms of the single ventricle, such as those associated with the asplenia and polysplenia syndromes and mitral atresia (16).

Periodic follow-up of operative survivors, in many now 10 or more years after the Fontan procedure (15,17,18), reveals that most are active and leading quality lives. Although late complications can occur, the most troublesome being atrial dysrhythmias (19), protein-losing enteropathy (7) and late ventricular deterioration and failure (15,17), there is accumulating evidence that evolving surgical techniques, such as the lateral tunnel approach (6) and the move to perform the Fontan procedure in the first 2 to 3 years of life, before the ventricle is exposed to a prolonged period of volume overloading and hypoxemia, will lead to continuously improving late results (14). Many female patients who have had the Fontan procedure now are reaching childbearing age, and although to date the total experience is small, a recent review (20) indicates that women who have had a good clinical result have an excellent chance of successfully completing a pregnancy and delivering without compromise of their cardiovascular status.

Choice of definitive surgical procedure. Pulmonary atresia with an intact ventricular septum is characterized by marked morphologic heterogeneity (12). It has been well established, however, that the size of the tricuspid valve and the size of the right ventricular cavity have a strong direct correlation (4). In patients with only mild to moderate tricuspid valve and right ventricular hypoplasia (Fig. 1) with a well-developed infundibular portion of the right ventricle (tripartite ventricle) and no coronary artery abnormalities, the neonatal surgical strategy is clearly to perform a right ventricular outflow tract reconstruction (2,4), and we favor concurrently performing a systemic to pulmonary artery shunt. This achieves the goal of establishing adequate pulmonary blood flow, and the forward flow through the decompressed ventricle encourages further growth of the tricuspid annulus and right ventricular cavity (21). Satisfactory palliation is achieved, and often definitive surgical correction, which involves closure of the atrial septal defect and division of the shunt, establishing a two-ventricle repair, can be accomplished in the first year or two of life (2,4).

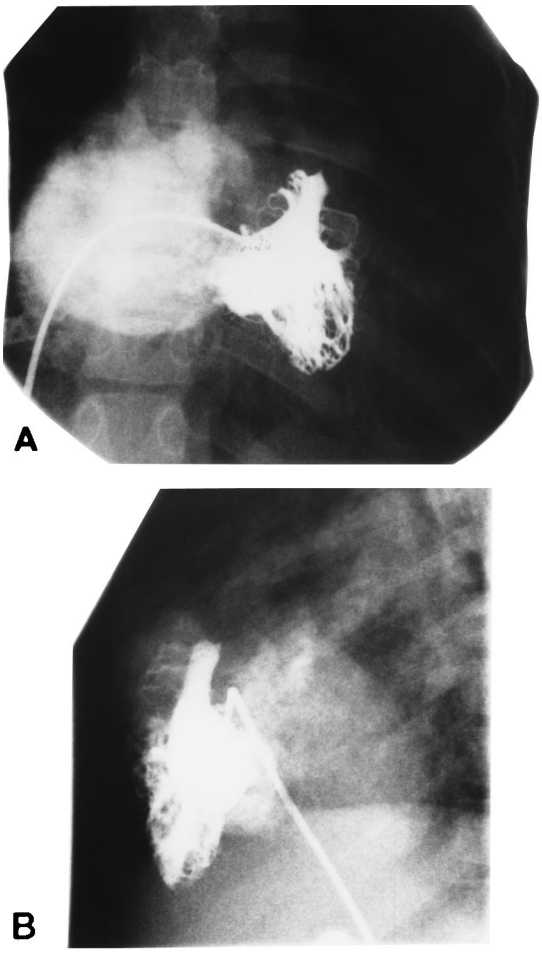

Figure 1. A, Angiocardiogram showing pulmonary atresia with an intact ventricular septum and moderate tricuspid valve and right ventricular hypoplasia. This neonate's tricuspid valve diameter measured 60% of that expected, and his tripartite right ventricular volume was 66% of that expected. Right ventricular outflow tract reconstruction and a Blalock-Taussig shunt were performed, and at age 18 months tricuspid valve diameter and right ventricular volume were both ~90% of that expected, permitting a two-ventricle complete repair at that time. **B,** Lateral view.

For this two-ventricle repair to be accomplished, we believe that both the tricuspid valve diameter and the right ventricular volume must be a minimum of 70% of that expected for the patient's body surface area. This determination is made angiographically, with additional echocardiographic assistance if necessary. If the tricuspid valve diameter and right ventricular volume are borderline for a two-ventricle repair, the repair is attempted, and if intraoperative postrepair right heart pressures and cardiac output are unsatisfactory, cardiopulmonary bypass is reinstituted and a bidirectional cavopulmonary anastomosis is performed, channeling superior vena caval blood directly to the pulmonary arteries while inferior vena caval flow continues to pass through the tricuspid valve and right ventricle (the so-called ventricle and one-half repair). In a like manner, we believe that the tricuspid valve diameter and right ventricular volume must be at least 50% of the expected size to

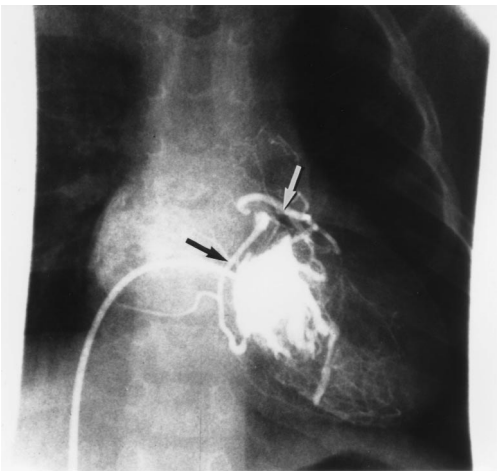

Figure 2. Angiocardiogram showing pulmonary atresia with an intact ventricular septum and a very hypoplastic tricuspid valve and unipartite right ventricle. Fistulous connections between the right ventricle and coronary arteries (**arrows**) are apparent. This diminutive tricuspid valve and right ventricle do not have significant growth potential, and this patient's most definitive surgical option is a Fontan procedure.

permit this "ventricle and one-half" type of repair. If the result of preoperative angiographic and echocardiographic assessment is in this range, this repair is done and, once again, intraoperative postrepair pressures and cardiac output are determined. If the hemodynamic data are unsatisfactory, bypass is reinstituted and a lateral tunnel pathway, directing inferior vena caval blood to the pulmonary arteries, is constructed (6), completing the Fontan circulation.

In retrospect, three of our patients in the Fontan series presented in this review had a tricuspid valve and right ventricular size that today may well have made them satisfactory candidates for the "ventricle and one-half" repair. However, they had their Fontan procedure performed before the concept of the bidirectional cavopulmonary anastomosis had evolved and were clearly not candidates for a two-ventricle repair, and therefore underwent the Fontan procedure.

Right ventricle to coronary artery fistulas. The presence of right ventricle to coronary artery fistulas can complicate surgical management, particularly if accompanying obstructive coronary lesions make portions of the myocardium dependent on the right ventricle (4,11,12,22). Freedom et al. (23,24) reported a 20% to 25% incidence of such right ventricular coronary dependency in patients with this lesion at their institution. These fistulas occur most frequently in patients with a very hypoplastic tricuspid valve and right ventricle, patients who are Fontan candidates only (14) (Fig. 2). If no right ventricular coronary dependence is produced by obstructive coronary artery lesions, the lateral tunnel Fontan procedure is done, and some centers (12,22) have recommended that the hypoplastic right ventricle be obliterated at the time of the procedure to reduce the risk of coronary artery "steal" (11) into the right ventricle through these fistulas with secondary myocardial ischemia. If there is right ventricular coronary dependence, the right ventricle is not obliterated and the atrial septal defect is left open, routing fully saturated pulmonary venous blood to the right ventricle and right ventricle-dependent parts of the coronary circulation and the desaturated inferior vena caval blood on the lateral side of the lateral tunnel patch within the atrium to the pulmonary arteries.

On rare occasions, a patient with a large tricuspid valve and right ventricle may have right ventricle to coronary artery fistulas, and if there are also obstructive coronary artery lesions producing right ventricular coronary dependence for a portion of the myocardium, a two-ventricle repair, which would decompress the right ventricle, is not possible. One of the patients in our current series had such an anatomy, and the Fontan procedure was performed with the lateral tunnel method and the atrial septal defect was left open so that pulmonary venous blood reached the right ventricle and right ventricle-dependent portions of the coronary tree.

Favorable selection bias in this series. One final comment must be made regarding the early and late results achieved with the Fontan procedure in this group of patients with pulmonary atresia and an intact ventricular septum. These patients were obviously "survivors" of the well-known treacherous early course that infants with this lesion may follow (3), all of them having been referred to our institution for operation at nearly 2 years of age or older. The finding of right ventricle to coronary artery fistulas in only five of these patients, an incidence considered to be lower than that usually found in this lesion as a whole (10,11), is further evidence that this group of patients was selectively a particularly favorable one. Thus, the relatively low operative mortality and late mortality in this series and the encouraging late follow-up status of survivors are probably better than those results that can be accomplished in a sequential series of patients presenting at a single institution as neonates (25). Nonetheless, the results should offer encouragement to patients with this lesion (and their parents), in whom the morphology of the tricuspid valve and right ventricle precludes a more corrective repair, leaving the Fontan approach as the best available option. As demonstrated in this series, such patients can have an excellent chance of surviving the operative procedure and then to continue to lead quality lives into adulthood.

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